

Enteric Duplication

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Abstract

Keywords

- ▶ alimentary tract duplication
- ▶ enteric duplication
- ▶ heterotopic mucosa
- ▶ colon
- ▶ rectum

Enteric duplications have been described throughout the entire gastrointestinal tract. The usual perinatal presentation is an abdominal mass. Duplications associated with the foregut have associated respiratory symptoms, whereas duplications in the midgut and hindgut can present with obstructive symptoms, perforation, nausea, emesis, hemorrhage, or be asymptomatic, and identified as an incidental finding. These are differentiated from other cystic lesions by the presence of a normal gastrointestinal mucosal epithelium. Enteric duplications are located on the mesenteric side of the native structures and are often singular with tubular or cystic characteristics. Management of enteric duplications often requires operative intervention with preservation of the native blood supply and intestine. These procedures are usually very well tolerated with low morbidity.

The term alimentary tract duplications was first described by Ladd in 1937. He was describing both the pathologic and clinical nature of these lesions as well as trying to unify multiple separate entities that had been previously described. Duplications are uncommon congenital lesions which can occur anywhere from the oropharynx to the anus.¹ The reported incidence of this is 1 in 4,500 births. Diagnosis and subsequent therapeutic intervention can be quite challenging based on the low frequency of these lesions. They are often found incidentally both in imaging studies and during other intraoperative explorations. The vast majority (70%) of patients present younger than 2 years with associated symptomatology which leads to early recognition and operative intervention. Currently, there does not appear to be any evidence that shows an association with sex, race, or genetic predisposition.²

There have been several different theories proposed about the etiology of interorgan locations. There does not appear to be a single unifying theory, however, that applies to the variety of duplications. However, there is usually a consistent mesenteric location of the duplications that are often lined with normal gastrointestinal epithelium.^{1,2} Some of the embryologic etiologies that have been described include environmental factors secondary to intrauterine vascular events or hypoxia, partial twinning, split notochord syndrome, remnants of embryologic diverticula, and abnormalities of recanalization.^{3,4}

In terms of location the vast majority are identified within the midgut specifically in the ileum. This is followed by the

esophagus, rectum, and colon. These are consistently on the mesenteric side of the bowel and are either directly adjacent or frequently form a part of the gastrointestinal wall. The blood supply to the duplication is usually shared in common with the native bowel. They are a true duplication often containing smooth muscle and gastrointestinal epithelium similar to the adjacent bowel wall.⁵

Upon resection and gross inspection, a single spherical duplication without communication to the adjacent bowel is often identified, whereas tubular duplications which are more commonly identified in communication. There have also been reports of heterotopic tissue identified within the duplications. The most common types of heterotopic tissue include gastric mucosal, squamous, transitional, columnar epithelium, pancreatic tissue, and lymphoid aggregates. Of these different types of heterotopic tissue the most significant of these is gastric and pancreatic which can be seen in up to a third of lesions and can result in hemorrhage, ulceration, and subsequent perforation.^{6,7}

Embryology

Briefly, the gut tube is formed from the endodermal lining of the yolk sac which becomes enveloped by mesodermal leading to the formation of the mesenteric vessels. The endoderm subsequently develops into the mucosal and submucosal structures. The mesoderm gives rise to the vascular supply as well as the

adventitia, serosa, lamina propria, muscularis mucosae, and the submucosal connective tissues. The enteric nervous system subsequently arises from migration of neural crest cells into the submucosa. Based on the arterial blood supply, the gut is divided into the foregut, midgut, and hindgut. The foregut is supplied by the celiac artery and its branches. The midgut and hindgut are supplied by the branches of the superior mesenteric artery and inferior mesenteric artery, respectively.^{8,9}

Alterations during normal embryological development have been attributed to subsequent gastrointestinal abnormality. Structural abnormalities attributed to embryologic maldevelopment include: esophageal atresia, midgut rotational abnormalities with peritoneal bands, mesenteric cysts, intestinal atresia, anorectal atresia, and enteric duplications. Specifically, for enteric duplications there have been several embryological pathways that have been studied. Of those there is some support for environmental factors, recanalization defects, split notochords, and partial twinning. Each of these theories suffers from not being able to identify a unifying cause.^{4,10,11}

Clinical Presentation

The emphasis on prenatal screening has led to the improved identification of anatomic pathology. Around 70% of patients are identified at younger than 2 years, secondary to early identification of abdominal masses as well as associated symptoms. As enteric duplications can be found anywhere along the alimentary tract, their presentation is often varied. Symptoms frequently overlap with other gastrointestinal abnormalities.³ In addition, many patients are asymptomatic. It is this population that is not identified in early childhood and is subsequently diagnosed into adulthood. Subsequent identification can often be an incidental finding during imaging obtained for workup of other intra-abdominal pathologies or trauma. Similarly, enteric duplications can be found during time of laparoscopy or laparotomy.¹²

Foregut duplications tend to present respiratory manifestations and with feeding intolerance.^{13,14} Midgut duplications are often associated with obstructive symptoms. Typical presentation includes feeding intolerance, early satiety, pain, nausea, emesis, and concern for an abdominal mass. Depending on location, size, and body habitus these can frequently be palpated on abdominal examination. Mass effect on surrounding structures can also lead to obstruction of the vena cava, biliary tree, or ureters leading to hydronephrosis. Acutely, these patient can present to the emergency department due to secondary complications. Commonly this includes perforation, acute intestinal obstruction, volvulus, gastrointestinal bleeding, perforation, and a risk of malignant transformation in adulthood.^{15–17} Similarly, hindgut duplications may present with any of the above symptoms in addition to constipation, hematochezia, misdiagnosis as diverticular disease, and pelvic pressure, or discomfort.^{18,19}

Diagnosis

Outside the perinatal period, where some enteric duplications are identified on prenatal ultrasound, the clinical presentation

is variable. Rarely is there a palpable abdominal mass or any specific physical examination finding. However, rectal and anal duplications can often be identified through careful inspection of the perineum and digital rectal evaluation. The radiographic workup usually begins with plain films. While helpful in identification of foregut duplications as a mass lesion in the chest, they can be less definitive within the abdomen. Usually, plain imaging demonstrates a normal bowel gas pattern. Occasionally, there may be findings consistent with an obstructive pattern. Contrast studies, such as barium enemas may be helpful, especially if there is communication with the adjacent bowel. Other findings include external compression on the bowel or obstruction.²⁰

Ultrasonography is one of the most commonly used modalities that assist in the identification of enteric duplications. This usually identifies a well-defined, often singular anechoic mass. Ultrasound can also identify debris or intraluminal contrast. One of the other associated ultrasonographic findings is a double layered appearance of the mass. This is often referred to as the “gut wall signature.” This is usually not seen with other intra-abdominal cyst or abscesses.²¹ Cross-sectional imaging such as computed tomography or magnetic resonance imaging can also provide some additional information and demonstrate the relationship to the adjacent bowel. Furthermore, these modalities can aid in the identification of multiple duplications. In the absence of significant findings on imaging, diagnostic endoscopy, or laparoscopy can be a useful adjunct.²²

Management

Once the diagnosis has been made treatment is indicated to mitigate symptomatology and to prevent future or further complications. The mainstay of therapy is complete surgical resection. The approach is dictated based on the location of the duplication within the alimentary tract. The important principles are the identification and preservation of the blood supply to the native bowel. In addition to careful operative examination, preoperative imaging can help guide the resection. Resection can be performed through both traditional open approaches, as well as thoracoscopic, laparoscopic, and combined with endoscopy.²³

For the purpose of this article the approach to foregut and midgut duplications will be briefly reviewed. Resection of duplications from the oropharynx to the thoracoabdominal region are typically approached from the right side via thoracoscopy or thoracotomy. Duplications that are associated with the diaphragm often require a thoracoabdominal approach through separate incisions for successful excision. The use of a transorally placed bougie can help guide the resection and leave an appropriately sized conduit (**►Fig. 1A–C**). Similarly, gastric, and duodenal duplications are amenable to complete resections. The caveat to this is the proximity of the duplication to the pancreaticobiliary tree. Intraoperative cholangiogram and ultrasound are helpful adjuncts to determine the extent of resection. In rare instances when the biliary tree shares the blood supply with the duplication, complete resection may not be possible. In this case partial resections with mucosectomy or internal drainage via cystenterostomy is appropriate. The remainder

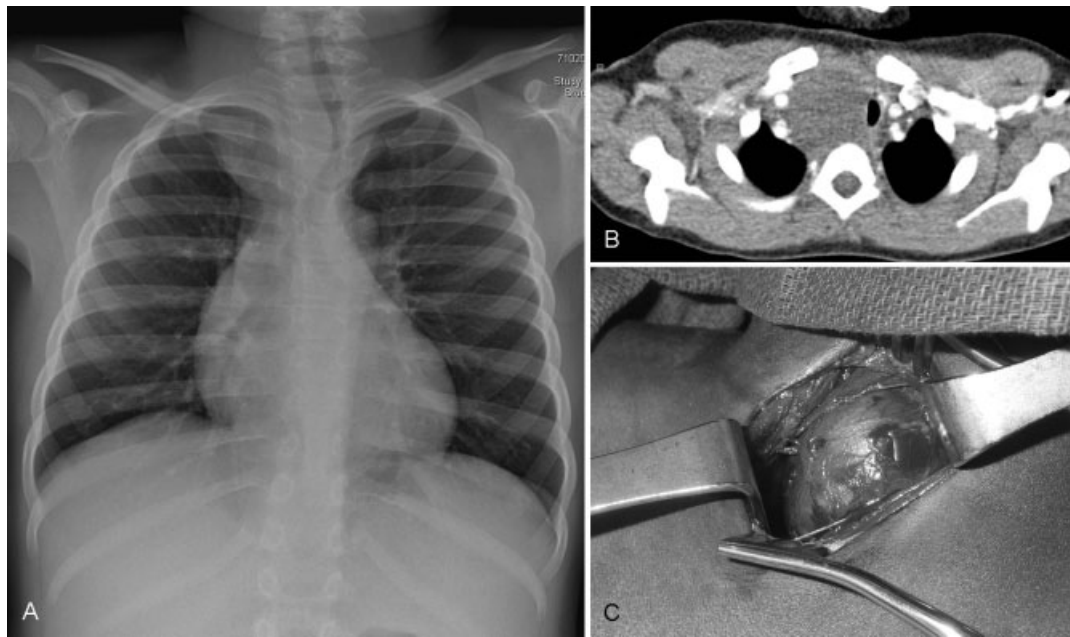


Fig. 1 Foregut duplication. (A) Plain radiograph showing a mass lesion in the upper chest—note the tracheal deviation. (B) Representative cross-sectional image showing an esophageal duplication cyst abutting the native esophagus into the right chest. (C) View of the esophageal duplication as seen through a cervical incision.

of the small intestine, which is the most common site for duplications, is usually resected with primary anastomosis. Cystic lesions are most amenable to this approach (► **Fig. 2**). More extensive, tubular lesions, require a combination of partial resection and mucosectomy to remove any heterotopic gastric mucosa.^{2,24,25}

Enteric duplications of the hindgut are often extensive and can be associated with abnormalities of the genitourinary system. In addition, while cystic and singular duplications can often be easily excised, tubular duplications are often more complex and may require a subtotal colectomy (► **Fig. 3**). Preoperative evaluation via endoscopy and cystoscopy are invaluable in providing additional information about this complex anatomy.²⁴ In addition, there has been a recent report of adenocarcinoma diagnosed within a cecal duplications. Management required adherence to standard oncologic principles of

resection as well as appropriate chemotherapeutic intervention. The presence of an elevated carcinoembryonic antigen in conjunction with a colonic duplication is highly suspicious for the presence of malignancy.²⁶

Rectal duplications are often asymptomatic until adulthood, where they are often diagnosed secondary to pain, hematochezia, or constipation. These can often be readily identified through cross-sectional imaging (► **Fig. 4A, B**). Several different approaches have been described for management of rectal duplications. The parasacral approach has the benefit of avoiding a laparotomy incision, and can be performed with low morbidity.²⁴ Laparoscopic resections, especially for large lesions, can often be safely performed with good results.²⁷ Advancements in transanal endoscopic



Fig. 2 Midgut duplication. At laparotomy, a singular cystic duplication is identified. This was subsequently managed with excision and primary anastomosis.

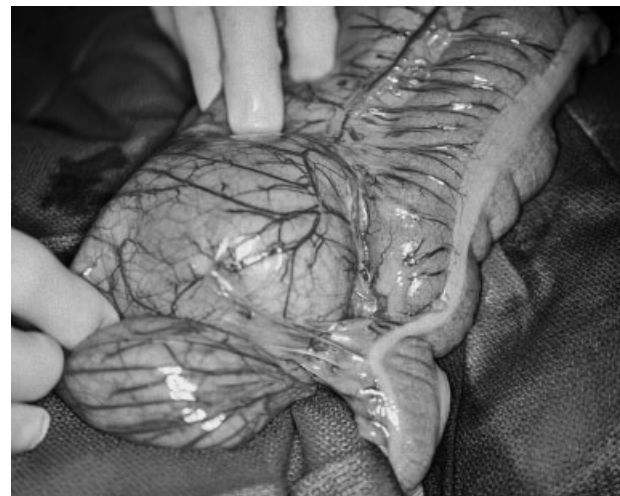


Fig. 3 Hindgut duplication, colon. A tubular duplication is seen in this representative photo taken at laparotomy.

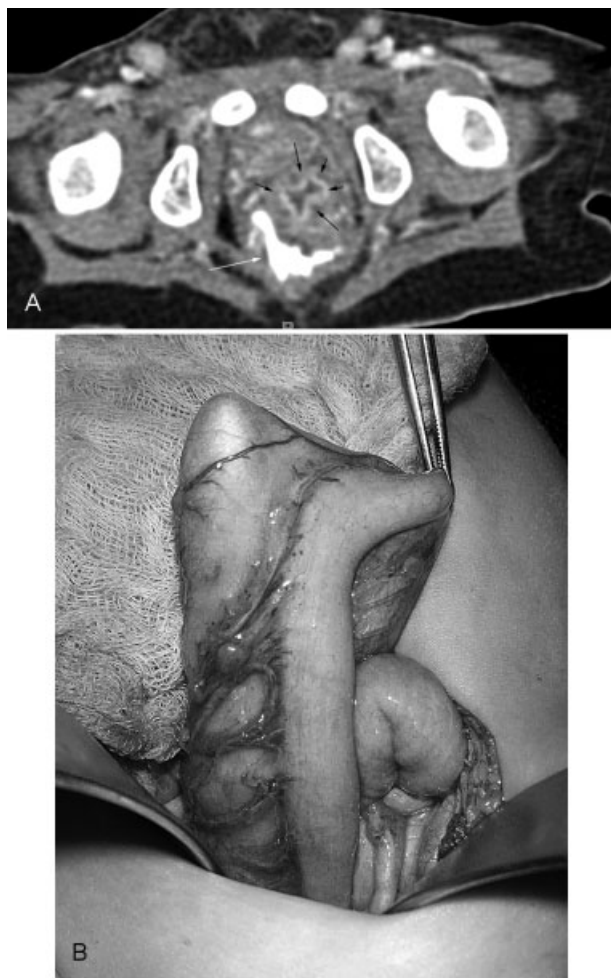


Fig. 4 Hindgut duplication, rectum. (A) Representative cross-sectional image of a rectal duplication as denoted by the black arrows. The contrast in the native rectum is seen as a separate structure (white arrow). (B) Tubular rectal duplication as seen at the time of laparotomy with dilator present within the lumen of the duplication. The shared blood supply is appreciated.

microsurgery have led to reports of successful resection through both combined and complete transanal approaches of small-to-moderate sized duplications.^{28,29}

Similar to the management of rectal duplications, anal duplications often involve associated anomalies of the genitourinary tract. Depending on the extent of the duplication, removal can be performed through a posterior sagittal or perineal approach with complete excision of the anal duplication.³⁰ In addition, there has been a case series of successful management through mucosectomy with primary repair avoiding unnecessary dissection.³¹

Conclusion

Enteric duplications are often identified before the age of 2 years secondary to a prenatal diagnosis of an abdominal mass or early symptomatology. Overall, these represent a rare portion of alimentary tract pathology. They can be found anywhere from the oral cavity to the anus. There is no unifying theory that encompasses the development of duplications, and

the current embryologic theories are heterogeneous. The vast majority are identified within the small bowel and are often associated with nonspecific clinical features. The overarching operative principles are the preservation of the adjacent vascular supply and the adjacent normal intestine, if at all possible.

While duplications of the colon, rectum, and anus are seen with less frequency, they are often more extensive and tubular in nature requiring resection and colocolostomy. Rectal duplications are often found in a presacral position without communication to the adjacent bowel. Excision can often be performed from a posterior sagittal approach avoiding entry into the adjacent rectum. A high index of suspicion needs to be maintained to diagnose an enteric duplication. Ultrasound has become an excellent modality to assist in the diagnosis. In the absence of other anatomic abnormalities, treatment of enteric duplications through operative resection is usually well tolerated, especially when diagnosis occurs into adulthood.

References

- Ladd WE, Gross RE. Surgical treatment of duplications of the alimentary tract enterogenous cysts, enteric cysts or ileum duplex. *Surg Gynecol Obstet* 1940;70:295–307
- Merrot T, Anastasescu R, Pankevych T, et al. Duodenal duplications. Clinical characteristics, embryological hypotheses, histological findings, treatment. *Eur J Pediatr Surg* 2006;16(01):18–23
- Ildstad ST, Tollerud DJ, Weiss RG, Ryan DP, McGowan MA, Martin LW. Duplications of the alimentary tract. Clinical characteristics, preferred treatment, and associated malformations. *Ann Surg* 1988;208(02):184–189
- Favara BE, Franciosi RA, Akers DR. Enteric duplications. Thirty-seven cases: a vascular theory of pathogenesis. *Am J Dis Child* 1971;122(06):501–506
- Macpherson RI. Gastrointestinal tract duplications: clinical, pathologic, etiologic, and radiologic considerations. *Radiographics* 1993;13(05):1063–1080
- Mellish RWP, Koop CE. Clinical manifestations of duplication of the bowel. *Pediatrics* 1961;27:397–407
- Puligandla PS, Nguyen LT, St-Vil D, et al. Gastrointestinal duplications. *J Pediatr Surg* 2003;38(05):740–744
- Soffers JH, Hikspoors JP, Mekonen HK, Koehler SE, Lamers WH. The growth pattern of the human intestine and its mesentery. *BMC Dev Biol* 2015;15(01):31
- Burns AJ, Roberts RR, Bornstein JC, Young HM. Development of the enteric nervous system and its role in intestinal motility during fetal and early postnatal stages. *Semin Pediatr Surg* 2009;18(04):196–205
- Bentley JFR, Smith JR. Developmental posterior enteric remnants and spinal malformations: the split notochord syndrome. *Arch Dis Child* 1960;35:76–86
- Bremer JL. Diverticula and duplications of the intestinal tract. *Arch Pathol* 1952;38:132–140
- Simsek A, Zeybek N, Yagci G, et al. Enteric and rectal duplications and duplication cysts in the adult. *ANZ J Surg* 2005;75(03):174–176
- Bratu I, Laberge JM, Flageole H, Bouchard S. Foregut duplications: is there an advantage to thoracoscopic resection? *J Pediatr Surg* 2005;40(01):138–141
- Lewis PL, Holder T, Feldman M. Duplication of the stomach. Report of a case and review of the English literature. *Arch Surg* 1961;82:634–640
- Jewett TC Jr, Walker AB, Cooney DR. A long-term follow-up on a duplication of the entire small intestine treated by gastroduplication. *J Pediatr Surg* 1983;18(02):185–188
- Orr MM, Edwards AJ. Neoplastic change in duplications of the alimentary tract. *Br J Surg* 1975;62(04):269–274

- 17 Ma H, Xiao W, Li J, Li Y. Clinical and pathological analysis of malignancies arising from alimentary tract duplications. *Surg Oncol* 2012;21(04):324–330
- 18 Fuchs JR, Clark K, Breckler FD, Rescorla FJ. Complete colonic duplication—a case report. *J Pediatr Surg* 2008;43(06):E11–E13
- 19 La Quaglia MP, Feins N, Eraklis A, Hendren WH. Rectal duplications. *J Pediatr Surg* 1990;25(09):980–984
- 20 Blickman JG, Rieu PH, Buonomo C, Hoogeveen YL, Boetes C. Colonic duplications: clinical presentation and radiologic features of five cases. *Eur J Radiol* 2006;59(01):14–19
- 21 Barr LL, Hayden CK Jr, Stansberry SD, Swischuk LE. Enteric duplication cysts in children: are their ultrasonographic wall characteristics diagnostic? *Pediatr Radiol* 1990;20(05):326–328
- 22 Hur J, Yoon CS, Kim MJ, Kim OH. Imaging features of gastrointestinal tract duplications in infants and children: from oesophagus to rectum. *Pediatr Radiol* 2007;37(07):691–699
- 23 Holcomb GW III, Gheissari A, O'Neill JA Jr, Shorter NA, Bishop HC. Surgical management of alimentary tract duplications. *Ann Surg* 1989;209(02):167–174
- 24 Hackam DJ, Zalev A, Burnstein M, Rotstein OD, Koo J. Enteric duplication in the adult, derived from the foregut, midgut and hindgut: presentation, patterns and literature review. *Can J Surg* 1997;40(02):129–133
- 25 Wrenn EL Jr. Tubular duplication of the small intestine. *Surgery* 1962;52:494–498
- 26 Jung KH, Jang SM, Joo YW, et al. Adenocarcinoma arising in a duplication of the cecum. *Korean J Intern Med* 2012;27(01):103–106
- 27 Salameh JR, Votanopoulos KI, Hilal RE, et al. Rectal duplication cyst in an adult: the laparoscopic approach. *J Laparoendosc Adv Surg Tech A* 2002;12(06):453–456
- 28 Rees CM, Woodward M, Grier D, Cusick E. Rectal duplication cyst: a combined abdominal and endoanal operative approach. *Pediatr Surg Int* 2007;23(04):369–371
- 29 Ben-Ishay O, Person B, Eran B, HersHKovitz D, Duek DS. Rectal duplication cyst in adults treated with transanal endoscopic microsurgery. *Tech Coloproctol* 2011;15(04):469–471
- 30 Jacquier C, Dobremez E, Piolat C, Dyon JF, Nagues F. Anal canal duplication in infants and children—a series of 6 cases. *Eur J Pediatr Surg* 2001;11(03):186–191
- 31 Tiryaki T, Senel E, Atayurt H. Anal canal duplication in children: a new technique. *Pediatr Surg Int* 2006;22(06):560–561